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Bilateral Sensorineural Deafness and Hydrocephalus Due to Foramen of Monro Obstruction in Sibs: A Newly Described Autosomal Recessive Disorder

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We identified a Canadian-Mennonite family in which a brother and sister have hydrocephalus due to obstruction at the foramen of Monro and profound bilateral sensorineural deafness. This appears to be a unique combination of anomalies and, to our knowledge, has not been reported previously. Both parents and a brother are phenotypically normal. The parents are second cousins. Thus, on the basis of consanguinity, affected sibs of both sexes, and in the absence of evidence for intrauterine infections or other adverse perinatal events, this syndrome is likely inherited in an autosomal recessive fashion. Am. J. Med. Genet. 68:350–356, 1997.

KEY WORDS: sensorineural deafness; hydrocephalus; autosomal recessive; foramen of Monro

INTRODUCTION

Approximately 1 in 1,000 children are born with bilateral sensorineural deafness, and over 60% are inherited [Morton, 1991; Reardon, 1992; Marazita et al., 1993]. Of those disorders known to be inherited, 75% are non-syndromic, with most inherited as autosomal recessive traits [Reardon and Pembrey, 1990; Marazita et al., 1993]. Autosomal dominant and X-linked traits also do occur but are less common. Gorlin et al. [1995] lists 22 non-syndromic genetic causes of hearing loss, whilst some estimate the number of loci to exceed 100 [Chung and Brown, 1970].

Several monogenic, non-syndromic causes of deafness have been mapped using DNA linkage analysis to various locations on the human genome; none have, to date, been cloned and sequenced. Some autosomal dominant forms have been localized to 5q31 (DFNA1) [Leon et al., 1992], 13q12 (DFNA2) [Chaïb et al., 1994], 1p32 (DFNA3) [Coucke et al., 1994], 19q13 (DFNA4) [Chen et al., 1995], 7p15 [Van Camp et al., 1995], and 4p16.3 (DFNA6) [Lesperance et al., 1995]. Genes for autosomal recessive deafness have been localized to 13q12 (DFNB1) [Chaïb et al., 1994; Guilford et al., 1994a; Scott et al., 1995; Brown et al., 1996], 11q13 (DFNB2) [Guilford et al., 1994b], 17q (DFNB3) [Friedman et al., 1995], 7q31 (DFNB4) [Baldwin et al., 1995], 14q (DFNB5) [Fukushima et al., 1995], 2p22-23 (DFNB6) [Chaïb et al., 1996], and 21q22 (DFNB8) [Veske et al., 1996]. There are other reports of autosomal loci mapped to different regions of the genome (P.J. McAlpine, personal communication). There appear to be at least two X-linked loci for deafness [Reardon and Pembrey, 1990]. One X-linked form of non-syndromic deafness has been localized to Xq21 [Bitner-Glindziez et al., 1994]. Half of the X-linked deafness families have an associated unique petrous temporal bone abnormalities with dilation of the internal auditory meatus and deficiency of the bone between the basal turn of the cochlea and the internal auditory meatus [Phelps et al., 1991].

Of the 25% which comprise the syndromic forms of deafness, Gorlin et al. [1995] lists 405 disorders, including a variety of chromosome syndromes, Usher syndrome, Waardenburg syndrome, (PAX3) [Tassabehji et al., 1993], Alport syndrome [Barker et al., 1990]. Not surprisingly, some mitochondrial mutations also lead to deafness [Van den Ouweland et al., 1994; Reid et al., 1994].

Congenital hydrocephalus occurs from between 0.4 and 0.9 per 1,000 livebirths [McCullough et al., 1983]. Non-neoplastic obstruction and/or aplasia of the fora-

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men of Monro is rare. This often presents as enlargement of the posterior horns of the parieto-occipital horns of the lateral ventricles or as unilateral or asymmetric hydrocephalus with normal-size third and fourth ventricles [Gaston and Jones, 1989; Wilberger et al., 1983]. Causes of obstruction at the foramen of Monro include thalamic tumors, abscesses, vascular anomalies, intrauterine infections, gliomatosis, ventricular diverticula, or congenital atresia [Oi and Matsumoto, 1985]. Hydrocephalus is highly heterogenous and can be due to single gene disorders such as X-linked hydrocephalus due to aqueduct stenosis or atresia, which is usually associated with other structural CNS abnormalities and mental retardation [Chow et al., 1985]. Hydrocephalus due to obstruction of the third ventricle is infrequent and is mainly due to lesions in the anterior portion, near the foramen of Monro. One family has been reported with two sibs with obstruction of the third ventricle and suspected autosomal recessive inheritance [Chow et al., 1990]. To our knowledge, there have been no reports of familial cases of foramen of Monro obstruction.

The occurrence of profound sensorineural hearing loss and obstruction of the foramen of Monro in two sibs of both sex, born to consanguineous and phenotypically normal parents, suggests a previously unrecognized autosomal recessive syndromic form of deafness.

CASE REPORTS AND INVESTIGATIONS

Case L.G.

This male child was the first born to a healthy 24-year-old mother and 25-year-old father who are second cousins (Fig. 1). The pregnancy was complicated by an upper respiratory tract infection and low grade fever at the time of conception. There was no exposure to known teratogens. He was born at term and weighed 4,010 g. Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. He was noted to have an enlarged head at birth (OFC = 43 cm) with no other defects or minor anomalies noted. A CT scan of the brain immediately following birth showed massive asymmetrical dilation of the temporal and occipital horns of the lateral ventricles, minimal dilatation of the frontal horns, and normal third and fourth ventricles, suggesting an obstruction at the foramen of Monro (Fig. 2a–c). An antibody screen for intrauterine infections and culture of urine for CMV was undertaken and results were normal. Ventriculo-

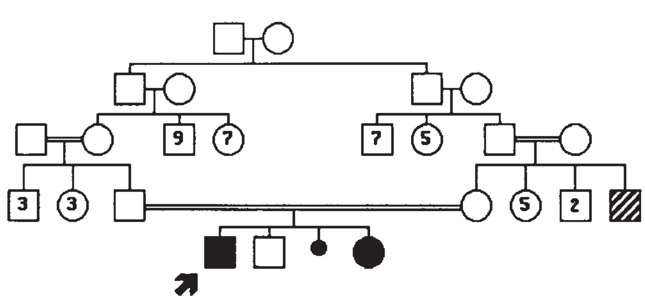

Fig. 1. Abbreviated pedigree of family. Solid symbols: affected children L.G. (propositus) and S.G. Hatched symbol: trisomy 21. Note consanguinity.

peritoneal shunts were placed in both ventricles in the first week of life.

His gross motor development progressed normally, but expressive and receptive language was delayed. An auditory brainstem response (ABR) assessment at age 2 months indicated normal hearing sensitivity bilaterally. A repeat ABR at 4 months again indicated normal hearing sensitivity bilaterally. Because of concerns of language delay, behavioral audiological testing was done at 22 months, which indicated a mild hearing loss in at least one ear. A third ABR was done at that time and responses were obtained to 30 dBnHL bilaterally, suggesting normal peripheral auditory function. Because of persisting language delay, visual alertness, and his attempt to communicate by gesture, behavioral audiological testing, and soundfield testing was repeated at 29 months and indicated a severe to profound sensorineural hearing loss bilaterally. L.G. was subsequently fit with binaural Unitron US80SA hearing aids and enrolled in an auditory-verbal clinic. His most current unaided audiological assessment took place at age 10.5 years and results indicated severe to profound hearing loss bilaterally. Aided testing indicated he receives good benefit from his hearing aids by reducing his thresholds to the mild hearing loss range. At age $10\frac{9}{12}$ years, ABR testing indicated no repeatable responses in neither ear at 95 dBnHL. These results were consistent with bilateral severe to profound hearing loss for the frequencies 2,000–4,000 Hz. Distortion Product Otoacoustic Emissions testing demonstrated no emissions bilaterally, which is consistent with bilateral hearing loss. Tympanometric measurements showed normal middle ear pressure with good tympanic membrane mobility bilaterally.

At 55 months his non-verbal development was in normal. His health has been good apart from the need to revise the V-P shunt on three separate occasions because of shunt blockage. He has developed an intermittent right exotropia, which was corrected with glasses. At the age of $9\frac{8}{12}$ years, he was attending a regular grade 4 class and doing age appropriate school work. Physical examination at that time showed a child with an essentially normal appearance (Fig. 3a,b) whose height was 126 cm (5–10th centile), weight was 24 kg (5th centile), and OFC was 53.6 cm (60th centile). He had downslanted palpebral fissures. His ears were mildly posteriorly angulated. He spoke with a slight hypernasal quality. No other abnormalities were detected.

Other investigations done during the course of evaluation of his hearing loss included normal visual evoked potential at 3 months, a normal ECG at 5 years, and normal chromosome analysis, 46,XY. A CT examination of the temporal and mastoid bones showed normal inner, middle, and external ear structures.

Case S.G.

This sister of L.G. was born at term following a normal and uncomplicated pregnancy, labour, and delivery. She weighed 3,950 g at birth. Apgar scores were normal. Her OFC at birth was 36 cm. She had mild elevation of serum bilirubin which was treated with phototherapy at 24–48 hours for 2 days. ABR assessments

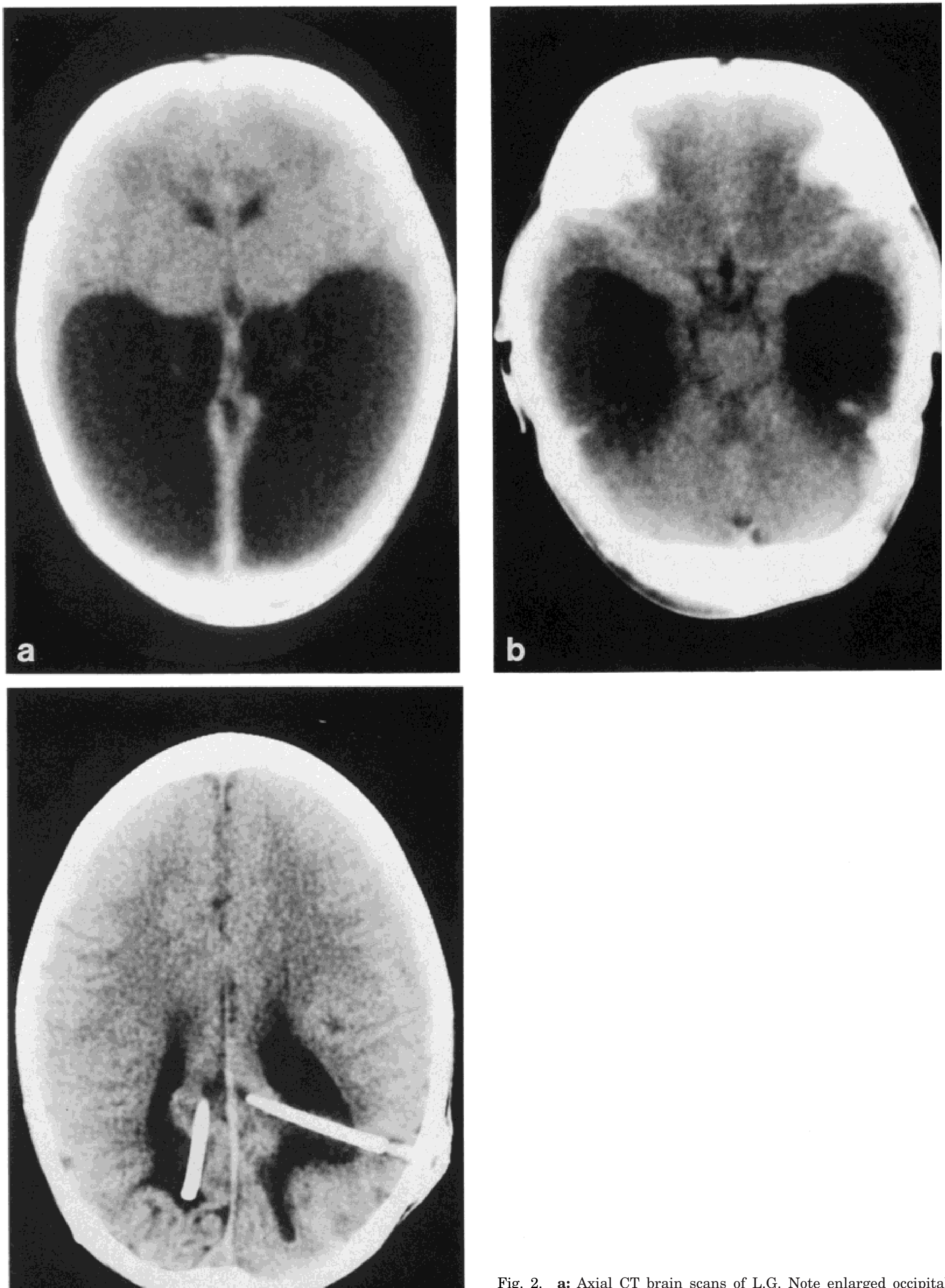

Fig. 2. **a:** Axial CT brain scans of L.G. Note enlarged occipital horns with mild asymmetry L > R, age 1 day. **b:** Note enlarged temporal horns and a normal third ventricle, age 1 day. **c:** Post shunting of both lateral ventricles. Note decompression and normal fourth ventricle, age 4 years.

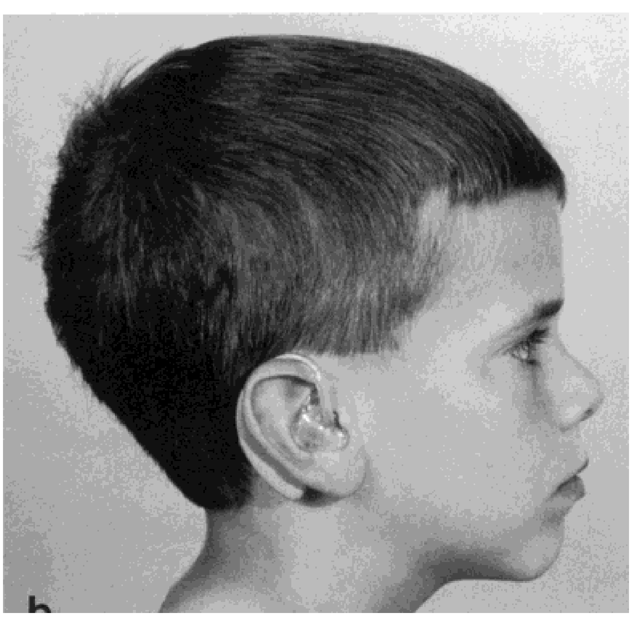

Fig. 3. Close up face (**a**) and profile (**b**) of L.G. at age 10 years. Note minimal dysmorphic features. Ears are mildly protruding, posteriorly rotated, and palpebral fissures are slightly down-slanted.

were carried out at 2 months and 3 months-of-age and indicated a severe to profound hearing loss bilaterally. She was subsequently fit with binaural Unitron US80PPL hearing aids and enrolled in an auditory-verbal clinic. Unaided audiological assessments indicated a severe to profound sensorineural hearing loss bilaterally. Aided testing at 29 months indicated that she received excellent benefit from her amplification by reducing her threshold to the mild hearing loss range. At 35 months, ABR testing showed no repeatable responses in either ear at 95 dBnHL, which is consistent with a bilateral severe to profound hearing loss for the frequencies 2,000 Hz–4,000 Hz. Distortion Product Otoacoustic Emissions testing demonstrated no emissions bilaterally, which is consistent with a bilateral hearing loss. Tympanometric measures revealed normal middle ear pressure with good tympanic membrane mobility bilaterally.

At 5 months her growth parameters showed her weight was 7.9 kg (90th centile), length was 67 cm (75th centile), and OFC was 45 cm (>95th centile). She had a prominent forehead, apparent hypertelorism and posteriorly angulated ears that were normally formed (Fig. 4a,b). The anterior fontanelle was full. Apart from this, the physical findings were normal. Development had progressed normally.

A cranial ultrasound at 9 months showed marked dilatation of the right lateral ventricles. A brain CT scan at 13 months showed massive dilatation of the right lateral ventricle, particularly of the temporal and occipital horns. The left lateral ventricle was mildly dilated, with normal third and fourth ventricles (Fig. 5a,b), which suggested partial obstruction of the foramen of Monro.

At 15 months, a neurosurgical consultation was obtained and she underwent a craniectomy with neuroendoscopic opening of the foramen of Monro and fenestration of the interventricular septum. During endoscopy, a thin veil of tissue was observed to be occluding the foramen of Monro on the right side. The tissue was opened with biopsy forceps and the septum pellucidum was fenestrated to allow communication between both lateral ventricles. Following the procedure, there was no obvious progression of the hydrocephalus, although the brain CT scan findings remained the same immediately postoperatively and 2 years later. Further neurosurgical treatment was considered unnecessary at that time. At the most recent follow up at 3 years, the child was developmentally and neurologically normal.

Other investigations done during the course of evaluation showed that she had a normal ECG, and CT examination of the temporal and mastoid bones showed normal inner, middle, and external ear structures.

DISCUSSION

Both sibs have bilateral profound sensorineural deafness. The oldest, L.G., was suspected to have a hearing loss from an early age in spite of three normal ABR assessments. This may reflect difficulty in interpreting ABR in younger children, or more likely, reflects a true progressive loss of hearing over the first 2 years of life. At 29 months, an unequivocally abnormal ABR was

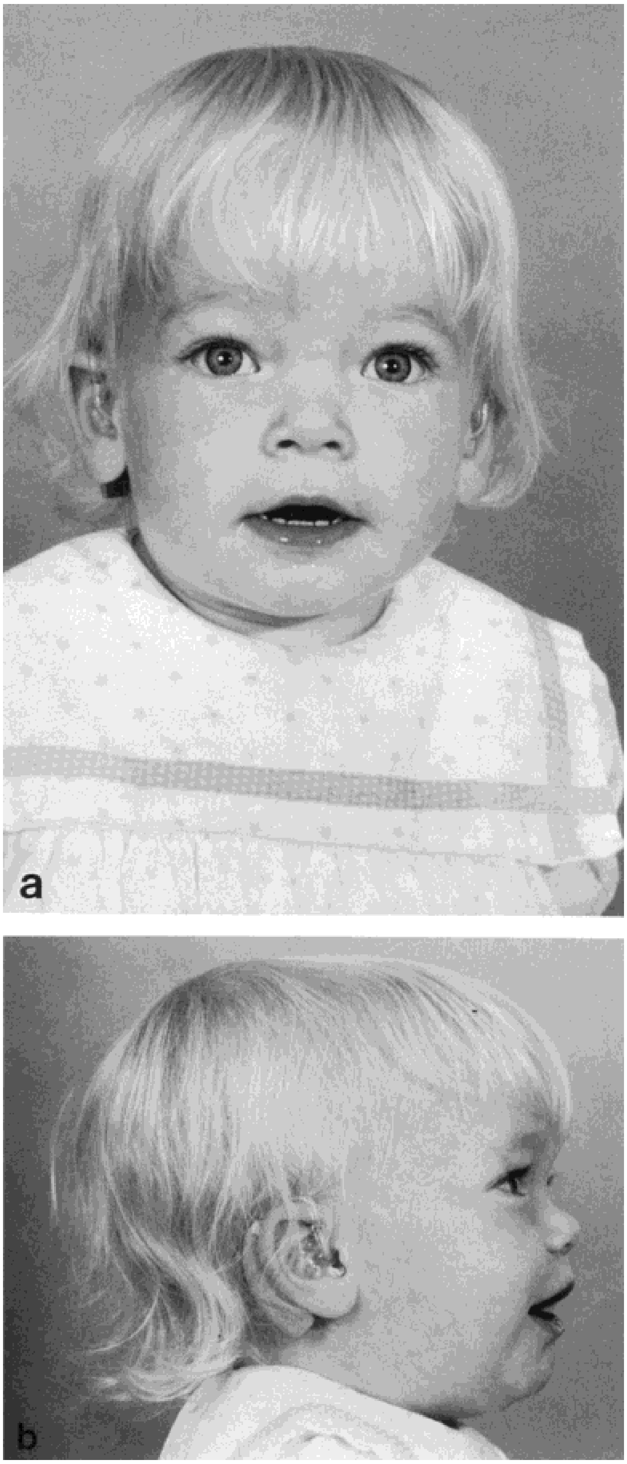

Fig. 4. Close up face (**a**) and profile (**b**) of S.G. at age 22 months. Note apparent hypertelorism and posteriorly rotated ears.

present and auditory amplification was prescribed with improvement in language and speech development.

The second affected sib was confirmed to have profound hearing loss at 2 and 3 months by ABR. Hearing was tested in her because of the unexplained hearing loss in her oldest brother, and because of our suspicion of hereditary deafness in light of the parental consanguinity. Neither child had external phenotypic abnormalities apart from the hydrocephalus. Their unaffected brother and both parents have normal hearing and appear phenotypically normal without evidence of hydrocephalus.

Familial hydrocephalus due to obstruction of the third ventricle has been reported once [Chow et al., 1990]. They described two sibs, one of whom died at 10 weeks of age. The other child was neurologically and developmentally normal with intact hearing. They suggested this was due to an autosomal recessive gene. Our two cases had no evidence of third ventricle obstruction. The marked asymmetrical dilatation of the lateral ventricles in S.G., and the normal-sized third and fourth ventricles in both S.G. and L.G. strongly support obstruction at the foramen of Monro.

Taboada et al. [1979] reported on two unrelated newborn infants who presented with asymmetrical hydrocephalus due to congenital atresia of the foramen of Monro. The family histories in those children were unremarkable. Unfortunately, the authors did not address the hearing status of the children. These authors reviewed other reports, which were scant in number, and all appeared to be sporadic in occurrence.

The mechanism for obstruction of the foramen of Monro is uncertain. It might be due to focal destruction or abnormal development of the ependyma with resulting collapse of one or both lumena as suggested by suckling hamsters and mice exposed to myxovirus [Johnson and Johnson, 1969]. Other pathophysiologic mechanisms of foramen of Monro obstruction were thoroughly reviewed by Oi and Matsumoto [1985].

In L.G., the hydrocephalus appears to be due to a partial obstruction of both foramena and complete obstruction on one side in S.G. with partial obstruction on the contralateral side. Regardless, the surgical treatment for obstruction and/or atresia of both foramen of Monro usually requires shunting of both lateral ventricles. To date, L.G. has not required any shunt procedures.

Many genetic causes of non-neural tube related hydrocephalus are associated with multiple, severe developmental anomalies of the brain and severe mental and physical handicaps. The two affected children in our report show no significant developmental delays, suggesting that the presence of this particular form of hydrocephalus can be associated with a normal neurodevelopmental outcome.

We undertook an extensive review of the literature in search of other reports of families with foramen of Monro obstruction and sensorineural deafness. None of the sources consulted revealed any such cases [McKusick, 1994; POSSUM, 1994; Gorlin et al., 1995]. Thus, we conclude that this family represents a newly described profound sensorineural hearing loss/hydrocephalus syndrome, likely due to autosomal recessive inheritance. Although there is a chance that these children are homozygous for two independent recessive traits, or remotely due to an unbalanced segregation

Fig. 5. **a:** Axial CT brain scans of S.G. Note marked asymmetry and unilateral hydrocephalus. Marked dilation of right occipital horn and mild dilation of left occipital and right frontal horns. Age 13 months. **b:** Right temporal horn dilatation and normal fourth ventricle. Age 13 months.

product of a cryptic chromosome translocation, the most parsimonious explanation is that these anomalies are due to the homozygous state of a single autosomal recessive gene.

It would be of interest, and we plan to pursue DNA linkage studies in this family to determine if the gene responsible maps close to loci identified in other autosomal recessive sensorineural deafness families. This might clarify inheritance as being autosomal recessive and might exclude other etiologies.

Finally, we recommend that hearing be carefully and serially assessed in children who present with hydrocephalus, which is suspected to be due to foramen of Monro obstruction. Conversely, children with sensorineural hearing loss and macrocephaly should have appropriate neuroradiologic studies.

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